Twin reverse arterial perfusion sequence

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ABSTRACT

Twin reverse arterial perfusion sequence occurs in approximately one percent of monochorionic twins. This condition is always fatal for the recipient twin and carries a high mortality rate for the pump twin. Various treatment options are described, but management is continually evolving with the publication of new data. We report an acardiac acephalic monochorionic twin who was diagnosed at 31 weeks gestation. Serial ultrasonographical examinations of the normal pump twin showed intrauterine growth restriction but with no evidence of heart failure. A healthy pump twin was delivered by caesarean section at 34 weeks.

Keywords: acardiac twin, intrafoetal ablation, monochorionic twins, twin reverse arterial perfusion sequence

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INTRODUCTION

Acardiac twinning is a rare malformation characterised by a usually normal "pump" twin perfusing an anomalous "perfused" recipient sibling via an artery-to-artery anastomosis, with reversed direction of flow of arterial blood to the perfused foetus. This haemodynamic reversal is known as twin reversed arterial perfusion (TRAP). TRAP sequence can have hypoxic effects on the perfused foetus. The cephalic pole, most distal to the retrograde perfusion, is the most severely affected. Total or partial agenesis of the heart and acephaly can occur in the recipient twin, in addition to an absence of one or more anatomical structure.[1] The syndrome occurs in 1% of monochorionic multiple gestation or one in 35,000 births.[2] At least 20% of the monozygotic co-pump twins can be expected to have congenital anomalies. In fact, 55% of pump twins die in the newborn period.

CASE REPORT

A 28-year-old Malay woman, gravida 2, para 1, was referred at 29 weeks from a district hospital because of foetal abnormality of the second twin noted on ultrasonography (US). Detailed US at the foetomaternal unit, Hospital Universiti Sains Malaysia, at 31 weeks gestation, demonstrated a single placental mass, concordant gender, and thin intertwin membrane, which suggested a monochorionic diamniotic twin pregnancy. The second twin was noted to be abnormal. The first twin appeared ultrasonographically normal but with oligohydramnios. Estimated foetal weight was 1.3 kg.

The second twin had a breech presentation. The foetus had acephaly, there was no foetal heart, and multiple cysts were seen around the neck region. The upper spine was not well formed; there was increased upper body mass, and the arms appeared abnormal. The lower limbs and the lower part of the body were normal. There was no polyhydramnious present. US appearances were those of a monochorionic diamniotic twin pregnancy with acardia acephaly and multiple abnormalities of the co-twin. Doppler studies showed the pulsatility index and resistance index normal in the pump twin. The biophysical profile of the pump twin showed a score of eight reduced from ten, due to oligohydramnios.

Conservative management of the pump twin was decided. Two doses of dexamethasone were given to promote foetal lung maturity and the patient was seen by the neonatologist for counselling. Delivery by casearean section was planned at 34+ weeks gestation. It was considered that the increased upper body mass of the acardiac twin lying second, would obstruct labour. The patient had weekly serial Doppler US to monitor the progression of the twins. However, the patient was admitted to the labour ward at 34 weeks gestation in premature labour. The pump twin was leading in cephalic presentation. The abnormal second twin continued in a transverse lie. An uneventful emergency lower segment caesarean section was carried out under general anaesthesia with the easy delivery of the first twin. The second was delivered by breech extraction.

The pump twin, a baby girl, weighed 1.41 kg and had Apgar score of ten at one and five minutes. The baby was observed in the neonatal intensive care unit for four days. There was no evidence of heart failure and she was discharged well on day 23 of life.

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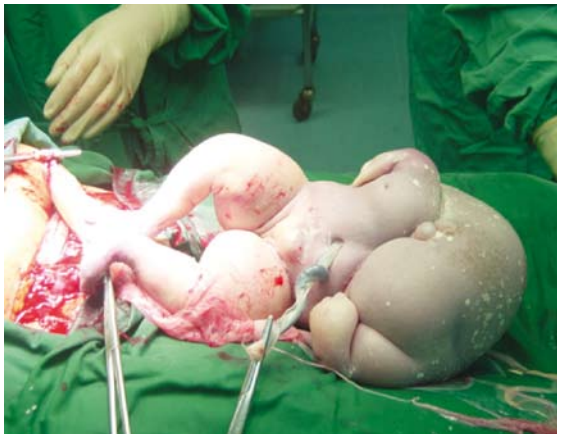

Fig. 1 Photograph shows the acardiac twin at delivery.

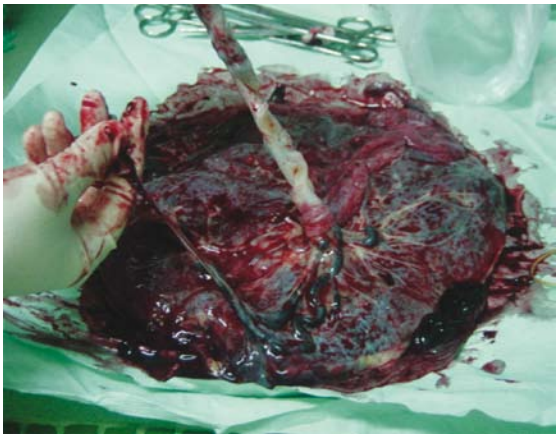

Fig. 2 Photograph shows the umbilical cord of the acardiac twin contains two vessels, one vein and one artery.

Inspection of the second twin, a female, who weighed 2.54 kg (Fig. 1), confirmed acephaly, an increased upper body mass which included the abnormal cephalic pole along with the abnormally-situated upper limbs and shoulders. Below the umbilical level, there was no obvious abnormality and the lower spine appeared normal. Autopsy was not possible nor were radiographs of the body obtained, as the father insisted on taking the baby and placenta immediately for burial. The intact placenta was monochorionic diamniotic in type and had demonstrable arterial and venous communications. The umbilical cord of the donor twin appeared normal, was 17 mm in diameter and contained three vessels, one vein and two arteries. The umbilical cord of the recipient twin was 6 mm in diameter, only one-third of the donor twin cord. It contained two vessels, one vein and one artery (Fig. 2).

DISCUSSION

Acardiac twin is a rare malformation resulting from extensive anastomoses between the placental vessels, with reversed flow TRAPS sequence in the recipient twin, causing hypoxia particularly to the cephalic pole leading to acardia acephaly and abnormalities of the upper body. The pump twin is at high risk, due to the hazards of congestive heart failure from a prolonged high output state plus the risk of prematurity. The mortality rate is reported to be between 50% and 70% for the pump twin.[2] Poor prognostic factors for the pump twin include acardiac-to-pump twin weight ratio of 70%, congestive heart failure and hydramnious in the pump twin.[2] Early US prenatal diagnosis of an acardiac foetus is problematic because the condition only becomes more pronounced in later pregnancy. Classically, as in this case, the Doppler US findings would then demonstrate the absence of cardiac movement, a reversed or abnormal blood flow through the umbilical cord, a poorly-defined head and trunk and abnormal upper limbs.

The optimal management of an acardiac twin pregnancy is controversial because of the rarity of its incidence and widely varied presentations, but the objective is clear: to achieve survival of the pump twin. The rate of foetal/neonatal mortality of the pump twin is high, therefore selected invasive procedures intended to deliver the acardiac twin[3] or to occlude blood flow to the acardiac twin have been practised by most authors. Such procedures include endoscopical umbilical cord ligation,[4] sclerosis of the umbilical cord with alcohol,[5] US-guided thermocoagulation of the umbilical cord and aorta,[6] endoscopical laser coagulation of the umbilical artery,[7] and bipolar coagulation of the umbilical cord.[8]

Quintero et al studied 65 patients with TRAP sequence and who were considered surgical candidates. Of these, 51 patients underwent umbilical cord occlusion. The overall perinatal survival for surgical candidates who had umbilical cord occlusion was 64.7% (33/51 patients) versus 42.9% (6/14 patients) for the surgical candidates who did not undergo umbilical cord occlusion.[9] They suggest that the surgical approach and surgical technique should be tailored to the specific clinical presentation, preferably by performing the surgery within the sac of the TRAP sequence foetus and avoiding disruption of the dividing membrane. Another prospective multicentre study by Hecher et al using foetoscopic laser coagulation of placental vascular anastomoses, or the umbilical cord of the 60 acardiac twin at median gestational age of 18.3 weeks, showed a survival rate of 80% and 67%, respectively, of

pregnancies with surviving pump twin going beyond 36 weeks of gestation.[10]

A new invasive method is by intrafoetal ablation of either the pelvic vessels or the abdominal aorta of the acardiac twin. These vessels are identified on colour Doppler US. Methods used in the intrafoetal ablation were by alcohol chemosclerosis,[11] monopolar diathermy,[12] interstitial laser[13] or radiofrequency.[14] A systematic review of minimally-invasive treatment modalities aiming to occlude vascular supply to acardiac twin has shown that the overall pump twin survival rate was 76%.[15] Intrafoetal ablation was associated with later median gestational age at delivery (37 versus 32 weeks) and higher median treatment-delivery interval (16 versus 9.5 weeks), compared with cord occlusion techniques. It was associated with a lower technical failure rate (13% versus 35%), lower rate of premature delivery or rupture of membranes before 32 weeks (23% versus 58%) and higher rate of clinical success (77% versus 50%) than cord occlusion techniques.[15]

A conservative approach has been described by Sullivan et al from ten cases of acardiac twins in which the neonatal mortality of the pump twin was less than reported from invasive management.[16] Nine of the ten women delivered healthy pump twins. Their management included two-weekly serial US, foetal echocardiography, Doppler flow study, along with performing a non-stress test and a biophysical profile. From the work of Sullivan et al, it would appear that conservative management with close antepartum surveillance in antenatally diagnosed TRAP sequences deserves consideration.[16]

The successful outcome in the case described in this paper is primarily related to the absence of structural heart defects, congestive heart failure, and hydramnios in the pump twin that survived. In conclusion, the management of the TRAP sequence still remains controversial. However, with the latest invasive interventions in treating the condition, there are hopes that the survival of the pump twin will be improved. These interventions should be considered and offered in parts of the world where interventional expertise is currently available.

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